

Trigeminal Neuralgia Diagnosis and Management – a Teaching Module

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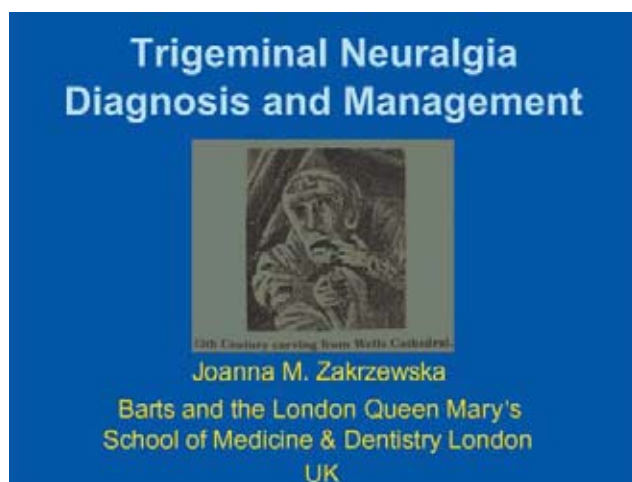
Summary: Below you will find a series of slides that were originally designed for an interactive Internet learning scheme, which provides an evidence based overview of the diagnosis and management of trigeminal neuralgia, a rare cause of facial pain. Although rare it is very important that any healthcare professional dealing with the face and mouth is able to recognize the condition as the management of this condition is very different from of the treatment of other orofacial pains. To make the most of this teaching aid the slides were designed to be viewed with the additional notes provided. The answers to the two exercises are provided at the end of the presentation. In addition you may wish to try the four questions listed below as part of your continuing professional development. The answers can be found at the end of this article.

Oral Biosci Med 2004; 1: 61-71

Submitted for publication 12 August, 2003; accepted for publication 1 October, 2003.

QUESTIONS

1. List the key features of trigeminal neuralgia.
2. How could you try to 'measure' pain in a way that you could apply to future evaluation of your treatment?
3. Mrs B is a fit 60-year-old woman with a 4-year history of classical trigeminal neuralgia. She has been pain free and off all medication for 6 months but in the last two weeks the pain has returned and is having a significant impact on her life.
 - a. What will you do in the short term?
 - b. What will you do in the long term?
 - c. How can you improve compliance with treatment?
4. Which drugs have been evaluated by randomised controlled trials and shown to be effective in the treatment of trigeminal neuralgia?



Slide 1



Slide 2



Diagnosis and Management can be improved by

- Use of a structured history
- Measurement of pain
- Determining level of distress
- Assessment of psychosocial factors
- Determining health beliefs/treatment goals



Slide 3

Trigeminal Neuralgia

IASP defines trigeminal neuralgia as:
 “a sudden, usually unilateral, severe, brief, stabbing, recurrent pain in the distribution of one or more branches of the fifth cranial nerve.”

Merskey et al, 1994.

Slide 4¹⁾

Exercise 1

Please read through the case studies in Table 2 and decide what you think is the possible diagnosis of the three cases?

- Trigeminal neuralgia
- Atypical trigeminal neuralgia
- Chronic idiopathic facial pain/atypical facial pain


Slide 5

International Headache Society Criteria for Trigeminal Neuralgia

A. Paroxysmal attacks of facial or frontal pain which last a few seconds to less than 2 minutes

B. Pain has at least 4 of the following characteristics:

1. Distribution along one or more divisions of the trigeminal nerve
2. Sudden, intense sharp, superficial, stabbing or burning in quality
3. Pain intensity severe



Slide 6²⁾

Trigeminal Neuralgia Criteria cont.


4. Precipitation from trigger areas, or by certain daily activities such as eating, talking, washing the face or cleaning the teeth

5. Between paroxysms the patient is entirely asymptomatic

C. No neurological deficit

D. Attacks are stereotyped in the individual patient

E. Exclusion of other causes of facial pain by history, physical examination and special investigation when necessary



Slide 7³⁾

Formal Measurements

McGill Pain Questionnaire (MPQ)

Visual Analogue Scales




Newton - John T. 2002

Slide 8⁴⁾

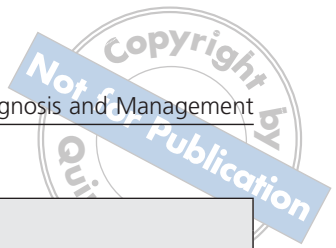


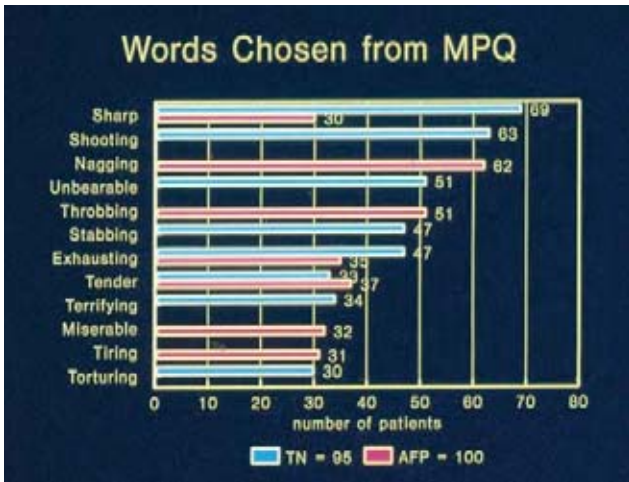
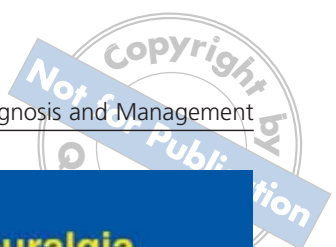
Table 1 McGill pain questionnaire

MCGILL PAIN QUESTIONNAIRE			
NAME:		DATE:	
Circle the word that describes how your pain feels right now:			
Nil			
Mild			
Moderate			
Severe			
Most Severe			
Circle the words below that best described your current pain:			
Use only <u>one word</u> in each group.			
Leave out any group if the words are unsuitable.			
1	2	3	4
Flickering	Jumping	Pricking	Sharp
Quivering	Flashing	Boring	Cutting
Pulsing	Shooting	Drilling	Lacerating
Throbbing		Stabbing	
Beating		Lancinating	
Pounding			
5	6	7	8
Pinching	Tugging	Hot	Tingling
Pressing	Pulling	Burning	Itchy
Gnawing	Wrenching	Scalding	Smarting
Cramping		Stinging	Stinging
Crushing			
9	10	11	12
Dull	Tender	Tiring	Sickening
Sore	Taut	Exhausting	Suffocating
Hurting	Rasping		
Aching	Splitting		
Heavy			
13	14	15	16
Fearful	Punishing	Wretched	Annoying
Frightful	Gruelling	Blinding	Troublesome
Terrifying	Cruel		Miserable
	Vicious		Intense
	Killing		Unbearable
17	18	19	20
Spreading	Tight	Cool	Nagging
Radiating	Numb	Cold	Nauseating
Penetrating	Drawing	Freezing	Agonizing
Piercing	Squeezing		Dreadful
	Tearing		Torturing

Table 2 Case studies for trigeminal neuralgia slide show

Please read these three case histories: do these patients have trigeminal neuralgia? How may you proceed to manage those with trigeminal neuralgia?

Case	1	2	3
Age	71	54	57
Gender	Female	Female	Female
Development of pain	Slowly developed first 5 years ago, there have been periods of weeks when there has been no pain	Constant over about 5 years, recent exacerbation which has not settled as usual, has not taken any medication for it as it was not "that bad".	First episode of pain 1.5 years ago beginning suddenly. Lasted for several weeks and then no pain for 9 months. Present episode of pain began 2 months ago after eating a meal.
Character/quality Words from McGill pain questionnaire	Quivering, jumping, pricking, sharp, gnawing, burning, stinging, aching, tender, tiring, wretched, annoying, piercing, numb, nagging	Aching heavy, nagging sometimes throbbing and sharp	Quivering, shooting, stabbing, sharp, crushing, tingling, aching, tender, tiring, terrifying, killing, blinding, unbearable, piercing, tight, agonizing
Site and radiation	Right mandible and nasolabial area, always the same area, felt deep in the face	Whole of the right side of the face	Left mental area is the trigger point and pain radiates up along the whole of the third division of the trigeminal nerve occasionally radiating to the outer canthus of the eye.
Severity Visual analogue scale 0–10 cm	At its worse 6 cm, mean of 3 cm, pain may go completely	Average 3.7 cm, at its worse 6 cm	Worse 9 cm, average of 4 cm, times when there is no pain.
Duration and periodicity	Each burst of pain lasts a few seconds these may repeat in episodes every few hours, periods of weeks of complete pain relief. Often left with a dull mild gnawing background pain.	Constant with intermittent severe episodes	Each pain episode lasts a few seconds but there may be bouts of these pains many times a day, may be no pain for a week or two. No pain at night
Provoking factors	Eating and brushing the teeth starts up pain	Opening mouth, eating, chewing and touching the area	Eating, talking, attempting to put make up on her lower lip, washing lower part of the face
Relieving factors	No activities	Nil	No activities
Associated factors	Some neck pain but no other pain or disturbances	Sometimes the area feels warm and appears reddened	Smoking makes the pain worse
Effect of pain on life style	Unable to socialise as much as would like, no evidence of anxiety or depression.	Divorced. Does not work, has had some impact on social life	Has a considerable effect on her quality of life, took a week of work as could not do her job as a personal secretary
Examination	No gross abnormalities, full denture wearer	No cranial nerve abnormalities and fully dentate with no dental disease	No cranial nerve abnormalities and fully dentate with no dental disease



Slide 9⁵⁾

Trigeminal neuralgia

Oh the horror of this pain
 The ice pick stabs again
 The quickness of the pain
 Has come and gone again
 Sometimes it lasts forever
 And I think that I might die
 At other times it is a burning
 Sometimes just a twitch
 My face may sometimes quiver

Slide 10⁶⁾

Exercise 2

- How would you manage the patient with trigeminal neuralgia ?
- Medical – which drug?
- Surgical – which surgery?

Justify your choices

Slide 11

Medical Management

Gold Standard – 3 RCTs
 Carbamazepine

- 70% of patients will respond with a reduction of pain
- may fail to offer relief because of increased severity of pain




Wiffen et al, 2002.

Slide 12⁷⁾

Carbamazepine (CBZ)

All patients will get side effects

- Drowsiness/tiredness
- Dizziness
- Zombie feeling
- Diplopia
- Ataxia
- Allergy 7%



Slide 13

Carbamazepine (CBZ)

- Liver enzyme inducer – raises gamma GT, liver enzymes
- Hyponatraemia at higher doses
- May depress white cell count especially initially
- Long term use may get depletion of folic acid
- Drug interactions – other anticonvulsants (AED), numerous others, warfarin

Slide 14

Baclofen

- Doses 40–80mg daily
- Can add to other AEDs
- Wide dosage range
- No major drug interactions
- Can use if patients allergic to CBZ
- Must increase and decrease drug slowly
- Use on three to four times daily regimen



Sindrup and Jensen, 1999.

Slide 15⁸⁾

Lamotrigine

- Doses 200–400mg daily
- Effective as add on therapy
- Can be used on regimen
- Safe in elderly
- Fewer drug interactions and side effects than CBZ
- Can rise dose rapidly due to rashes



Wiffen et al, 2002.
Zakrzewska and Lopez, 2003.

Slide 16⁹⁾

Other Drugs used in RCTs

- Pimozide – effective but severe side effects
- Tizanide – no better than cbz
- Tocainide – effective but severe adverse reactions
- Proparacaine eye drops – not effective

Sindrup and Jensen, 1999.
Zakrzewska and Lopez, 2003.

Slide 17¹⁰⁾

Phenytoin

- Doses up to 300mg daily
- Effectiveness reported in 29 cases
- Works synergistically with cbz
- Side effects and drug interactions common
- Need to replace folic acid in long term use



Slide 18¹¹⁾

Clonazepam

- Doses 2–8mg daily
- Effectiveness reported in 63 cases
- Lethargy, fatigue, dizziness very common and dose related
- Need to check regular liver function
- Must not be withdrawn rapidly



Slide 19¹²⁾

Gabapentin

- Doses 1200–3600mg daily
- Effectiveness in 33 cases although RCT in post-herpetic neuralgia effective
- Fewer side effects and drug interactions than cbz
- Need to start and withdraw slowly
- Expensive



Slide 20¹³⁾



Oxcarbazepine (OXC)

- Doses 300–1200mg daily
- OXC 300mg = CBZ 200mg
- Effectiveness shown in 53 cases
- Fewer side effects than cbz
- Hyponatremia dose related
- Fewer drug interactions than cbz

Zakrzewska and Patsalos, 2002.

Slide 21¹⁴⁾

PAIN DIARIES

Scoring system 0–5

Date	Pain intensity	Daily activities	Daily drug dose	Side effects	Stress /other
19.3	3	5	600mg cbz	Drowsy	3
20.3	3	4	600mg cbz	Drowsy	2

Slide 22¹⁵⁾

RCTs in Trigeminal Neuralgia

Drugs:	carbamazepine	effective
	lamotrigine	likely to be beneficial
	baclofen	likely to be beneficial
	pimozide	trade off benefit /harm
	tizanidine	unknown effectiveness
	propranolol	unlikely to be beneficial
	tocainide	harmful

Surgery:
peripheral streptomycin not beneficial

Zakrzewska and Lopez, 2003.

Slide 23¹⁶⁾

Surgical Management

Surgery becomes necessary when:

- Patients are no longer controlled with medications
- Side effects of medication are unacceptable
- Poor quality of life

Ablative techniques damage the nerve
Microvascular decompression is the only non ablative procedure
MRI may show evidence of compression
Timing of surgery remains unclear due to lack of evidence

Nurmikko and Eldridge, 2001.
Zakrzewska JM, 2002.

Slide 24¹⁷⁾

Peripheral Techniques

- Cryosurgery
- Neurectomy
- Alcohol injection
- Laser
- Acupuncture

Pain relief around 10 months

Slide 25¹⁸⁾

Gasserian Ganglion

- Radiofrequency thermorhizotomy (RFT)
- Percutaneous glycerol rhizotomy (PGR)
- Balloon microcompression

Slide 26¹⁹⁾

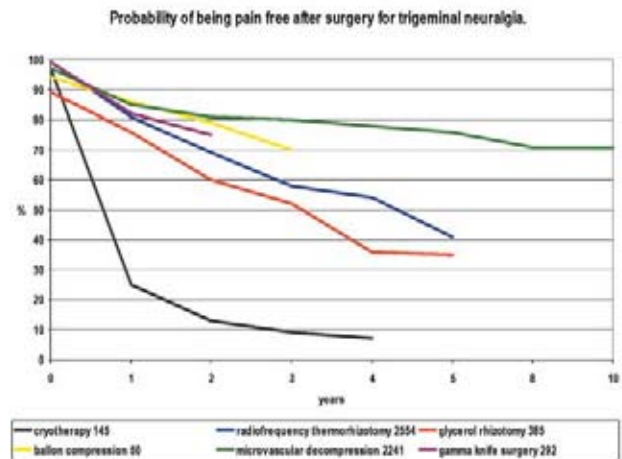


Posterior Fossa

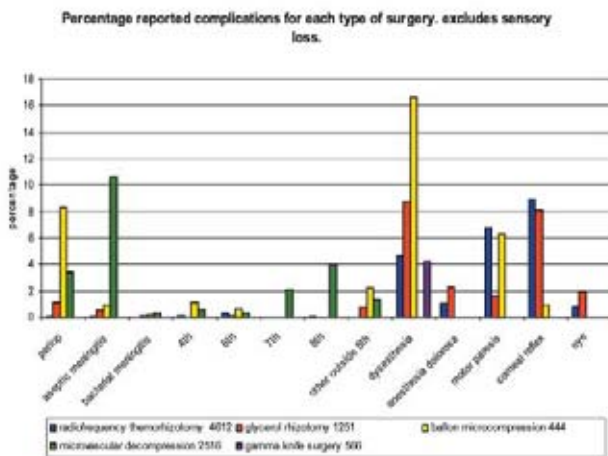
- Microvascular decompression (MVD)
- Partial sensory rhizotomy (PSR)
- Stereotactic radiosurgery - Gamma Knife Surgery (GKS)




Slide 27²⁰⁾



Slide 28²¹⁾



Slide 29²²⁾

Complications after Surgery

Mortality

- around 0.5% for MVD
- lower for all other procedures

Serious complications

most likely after MVD include intracranial haemorrhage and general medical complications as after any major surgery

Arrhythmias common with balloon microcompression

Slide 30²³⁾

Trigeminal Neuralgia

- Support groups in US and UK
- Web sites
 - www.tna-support.org
 - www.tna.org.uk
 - Telephone help lines
- Email/ chat rooms
- "Striking Back" – Weigel and Casey
- Brain and Spine Foundation Face Pain Booklet

Slide 31²⁴⁾

Answers to Exercise 1

- Case 1 – Atypical Trigeminal Neuralgia
- Case 2 – Chronic Idiopathic Facial Pain
- Case 3 – Trigeminal Neuralgia

Slide 32²⁵⁾

Answers to Exercise 2

- Start with cbz slowly increasing the dose
- If not well controlled on 800mg daily consider change of drug
- Do an MRI to assess compression

Slide 33²⁶⁾

ANSWERS

1. See slides 6-7
2. See slides 8-9
3.
 - a. If she has responded well to carbamazepine in the past start on this and work up to a dose around 800mg daily, give the patient a pain diary
 - b. Consider referral to a neurosurgeon for possible microvascular decompression as patient has classical trigeminal neuralgia and is medically fit. If the patient does not wish to have surgery but suffers side effects from medication may wish to change to other anticonvulsant therapy.
 - c. Provide more patient information, provide contact with support group involve patient in decision making.
4. Carbamazepine, baclofen, lamotrigine

NOTES

- ¹⁾ Merskey H, Bogduk N. Classification of chronic pain. Descriptors of chronic pain syndromes and definitions of pain terms. 2nd ed. Seattle: IASP Press 1994;59-60.
- ²⁾ These diagnostic criteria have not been validated by case control studies.
Olesen J. The international classification of headache disorders. 2nd edition. Cephalgia 2004;24:126-127.
Zakrzewska JM. Diagnosis and differential diagnosis of trigeminal neuralgia. Clin J Pain 2002;18:14-21.
Nurmikko TJ, Eldridge PR. Trigeminal neuralgia – pathophysiology, diagnosis and current treatment. Br J Anaesth 2001;87:117-132.
- ³⁾ Not all patients fit these criteria and some patients will also complain of aching, dull background pain that tends to be more continuous. This type of 'after pain' is often found in other neuropathic pain syndromes. When treated surgically

these patients often have some remaining dull pain as surgery seems to be mainly effective against sharp pain. This type of pain has been labelled as atypical trigeminal neuralgia. If this has been ascertained prior to surgery patients can be told that surgery may not give complete pain relief.

- ⁴⁾ The McGill Questionnaire text can be found in Table 1.
Newton-John T. Measurement of pain in adults in Assessment and management of orofacial pain. In: Zakrzewska JM, Harrison S (eds). Pain research and clinical management, volume 14. Amsterdam: Elsevier Sciences 2002;87-104.
- ⁵⁾ This study looked at 95 patients who fulfilled the criteria for trigeminal neuralgia with 100 who had atypical facial pain (AFP) or chronic idiopathic facial pain.
Zakrzewska JM. Trigeminal neuralgia. London: Saunders 1995; 78.
This was based on a study done by Melzack and Fromm.
Melzack R, Terrence C, Fromm G, Amsel R. Trigeminal neuralgia and atypical facial pain: use of the McGill Pain Questionnaire for discrimination and diagnosis. Pain 1986;27:297-302.
- ⁶⁾ This poem was written by a patient with trigeminal neuralgia.
- ⁷⁾ Wiffen P, McQuay H, Carroll D, Jadad A, Moore A. Anticonvulsant drugs for acute and chronic pain. Cochrane Database Syst Rev 2002; CD001133. (The Cochrane review is updated usually every two years)
Zakrzewska JM, Lopez BC. Trigeminal neuralgia. Clinical evidence concise 2003;9:280-281. (Latest version www.clinicalevidence.com)
- ⁸⁾ Sindrup SH, Jensen TS. Efficacy of pharmacological treatments of neuropathic pain: an update and effect related to mechanism of drug action. Pain 1999;83:89-400.
Zakrzewska JM, Lopez BC. Trigeminal neuralgia. Clinical evidence concise 2003;9:280-281. (Latest version www.clinicalevidence.com)
- ⁹⁾ Wiffen P, McQuay H, Carroll D, Jadad A, Moore A. Anticonvulsant drugs for acute and chronic pain. Cochrane Database Syst Rev 2002; CD001133. (The Cochrane review is updated usually every two years)
Zakrzewska JM, Lopez BC. Trigeminal neuralgia. Clinical evidence concise 2003;9:280-281. (Latest version www.clinicalevidence.com)
- ¹⁰⁾ The references provided have all the individual references to the original studies included in the reviews.
Sindrup SH, Jensen TS. Efficacy of pharmacological treatments of neuropathic pain: an update and effect related to mechanism of drug action. Pain 1999;83:89-400.
There is a regular annual update in the journal Clinical Evidence with the latest updates being available on its internet site.
Zakrzewska JM, Lopez BC. Trigeminal neuralgia. Clinical evidence concise 2003;9:280-281. (Latest version www.clinicalevidence.com)
- ¹¹⁾ No RCTs, case reports only.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- ¹²⁾ No RCTs.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- ¹³⁾ No RCTs.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Har-

- rison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- 14) No randomised controlled trial but there is a longitudinal cohort study.
Zakrzewska JM, Patsalos PN. Long term cohort study comparing medical (oxcarbazepine) and surgical management of intractable trigeminal neuralgia. *Pain* 2002;95:259-266.
- 15) These diaries are a good way of getting patients involved and in control of their pain. Patients can decide on their own format. For trials electronic diaries would be better.
- 16) Sindrup SH, Jensen TS. Efficacy of pharmacological treatments of neuropathic pain: an update and effect related to mechanism of drug action. *Pain* 1999;83:389-400.
Wiffen P, McQuay H, Carroll D, Jadad A, Moore A. Anticonvulsant drugs for acute and chronic pain. *Cochrane Database Syst Rev* 2000;CD001133.
Zakrzewska JM, Lopez BC. Trigeminal neuralgia. *Clinical evidence concise* 2003;9:280-281. (Latest version www.clinicalevidence.com)
- 17) There are few high quality trials of surgical management. As the trials are not randomised there is no way of knowing whether the surgery worked because of a natural period of pain remission.
Nurmikko TJ, Eldridge PR. Trigeminal neuralgia – pathophysiology, diagnosis and current treatment. *Br J Anaesth* 2001;87:117-132.
Zakrzewska JM, Lopez BC. Quality of papers reporting outcomes after surgical management of trigeminal neuralgia. Recommendations for future reports. *Neurosurgery* 2003;53:110-122.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
Zakrzewska JM. Diagnosis and differential diagnosis of trigeminal neuralgia. *Clin J Pain* 2002;18:14-21.
- 18) These techniques depend on localising a trigger spot and all can be done on an outpatient basis. Most are neuro-destructive and will cause local sensory loss.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- 19) Descriptions of these techniques can be found in a variety of textbooks and articles. These involve inserting a needle into the Gasserian ganglion and producing nerve damage.
Nurmikko TJ, Eldridge PR. Trigeminal neuralgia – pathophysiology, diagnosis and current treatment. *Br J Anaesth* 2001;87:117-132.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- 20) Descriptions of these techniques can be found in a variety of textbooks and articles.
MVD is a non ablative procedure which involves decompressing the trigeminal nerve usually caused by an artery. Partial rhizotomy is done when no compression is found and a part of the nerve is cut.
Gamma knife surgery is an ablative procedure which delivers radiation to the trigeminal nerve in the posterior fossa.
Nurmikko TJ, Eldridge PR. Trigeminal neuralgia – pathophysiology, diagnosis and current treatment. *Br J Anaesth* 2001;87:117-132.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- 21) Using only the top quality studies these Kaplan Meir survival curves show that microvascular decompression gives the longest period of pain relief – 70% at ten years will be pain free whereas ablative techniques provide 4-5 years of pain free time.
Lopez BC, Hamlyn PJ, Zakrzewska JM. Systematic Review of Ablative Neurosurgical Techniques in the Management of Trigeminal Neuralgia Neurosurgery. In press.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- 22) These complications show that peri-operative complications are highest after MVD and the commonest complication is loss of hearing around 4%. All other procedure result in complications related to trigeminal dysfunction – sensory loss in a variety of degrees.
Lopez BC, Hamlyn PJ, Zakrzewska JM. Systematic Review of Ablative Neurosurgical Techniques in the Management of Trigeminal Neuralgia Neurosurgery. In press.
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- 23) Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;267-370.
- 24) Patients greatly appreciate receiving more information as there is very little around. Both these support groups offer a range of materials and other countries have now set up groups e.g. Australia, and Spine Foundation Face Pain booklet ISBN 1 901893 16 2: 2002; or <http://www.brainandspine.org.uk>
Weigel G, Casey KF. Striking Back. The trigeminal neuralgia handbook. Barnegat Light: The Trigeminal Neuralgia Association 2000.
- 25) Case 1 – This patient also describes a dull type of pain which is more persistent and more in line with a neuropathic pain. It has been suggested that these patients show evidence of long term compression and nerve atrophy.
Case 2 – Although unilateral and with some sharp elements has an over riding constant pain.
Case 3 – This is a classical trigeminal neuralgia with all the diagnostic features as shown in slide 4.
- 26) An algorithm for management can be found in
Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD (eds). Assessment and management of orofacial pain. Amsterdam: Elsevier Sciences 2002;354.

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- Lopez BC, Hamlyn PJ, Zakrzewska JM. Systematic Review of Ablative Neurosurgical Techniques in the Management of Trigeminal Neuralgia Neurosurgery. In press.
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- Zakrzewska J.M, Patsalos PN. Long term cohort study comparing medical (oxcarbazepine) and surgical management of intractable trigeminal neuralgia Pain 2002;95:259-266.
- Zakrzewska JM, Lopez BC. Quality of papers reporting outcomes after surgical management of trigeminal neuralgia. Recommendations for future reports. Neurosurgery 2003;53:110-122.
- Zakrzewska JM, Lopez BC. Trigeminal neuralgia. Clinical evidence concise 2003;9:280-281. (Latest version www.clinicalevidence.com)
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